



Pulmonary Renal syndrome

Mohsen Elshafey

Pulmonary/Critical Care MD

Mansoura university

mohsenpccu@gmail.com

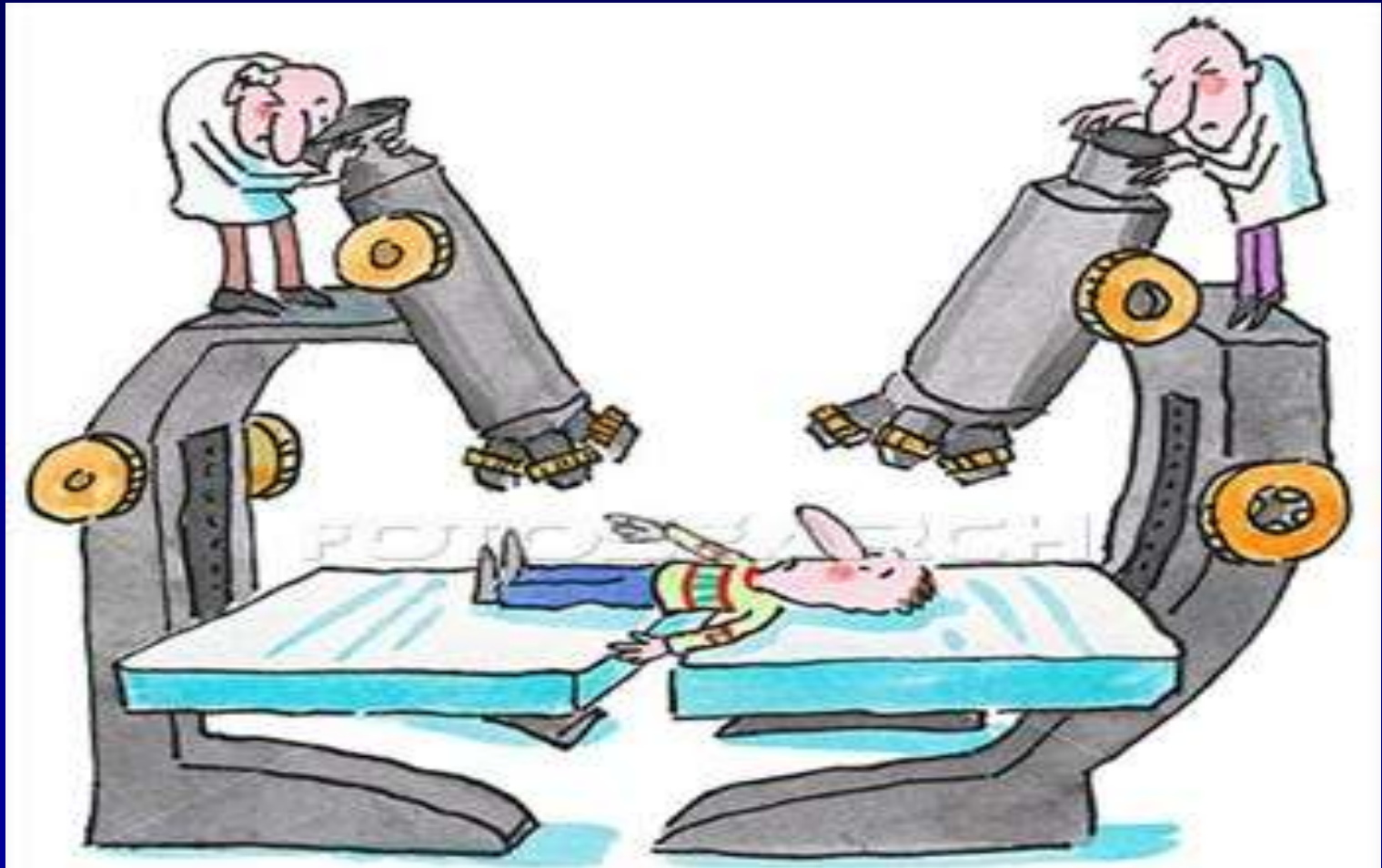
Kidney and lung in health and disease

- **Physiological : Acid/Base regulation .**
- **Pulmonary disorders affecting kidney.**
- **Renal disorders affecting lung.**
- **Pulmonary renal syndrom.**

Pulmonary manifestations of renal disorders

- **I**nfections.
- **H**ypoxemia.
- **E**ffusion.
- **P**ulmonary embolism.
- **P**ulmonary oedema
- **A**sthma.
- **P**ulmonary calcification / Infiltrations.
- **S**leep disordered breathing.

Pulmonary Renal Syndrome

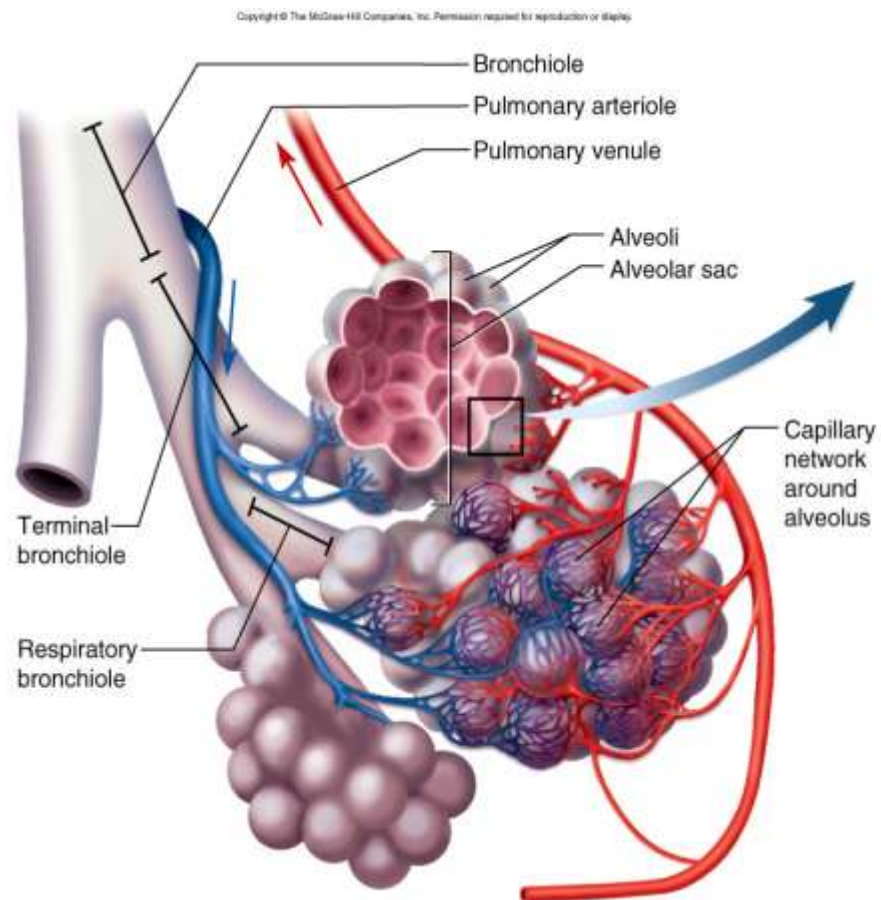
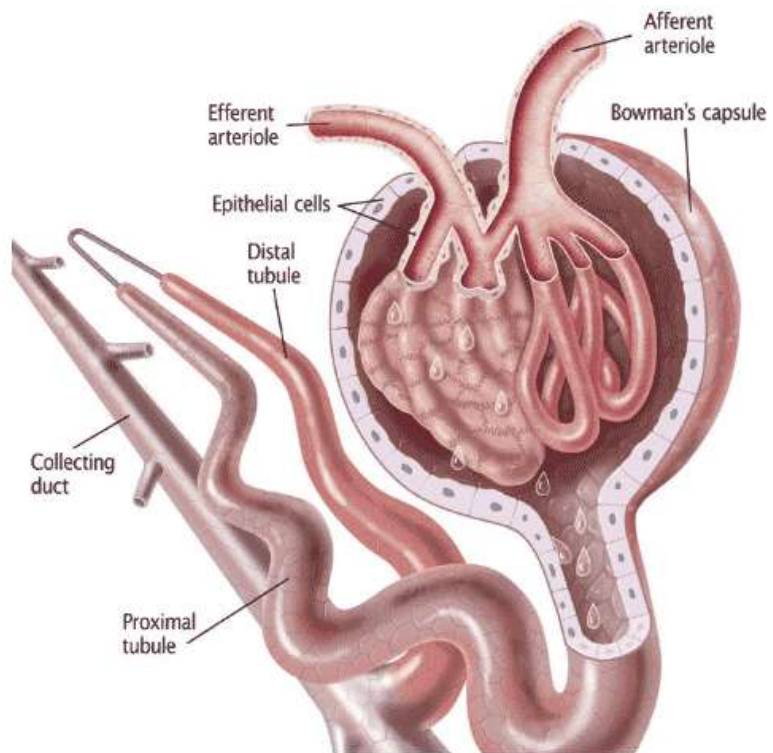


OBJECTIVES

- What based this syndrome .
- What is PRS. ?
- Aetiological /classification of PRS.?
- How can we diagnose.
- Therapeutic considerations.
- PRS from the word to ICU.



**What do the lung
and kidney have in
common?**





**Extensive amount of micro-vasculature
across a large surface area**



Basement membrane (special antigens)



**Exchange of materials across a thin
barrier**



**Both clean the body of waste
material and manage the delicate
balance of other materials.**



What is PRS

PRS is defined as the combination of:

- Diffuse alveolar hemorrhage (DAH)

and

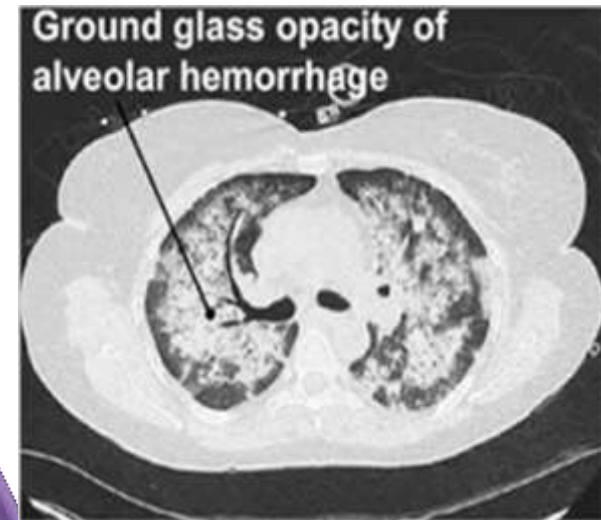
- Rapidly progressive glomerulonephritis.

1-DAH

Bleeding into the alveolar spaces due to disruption of the alveolar-capillary basement membranes caused by injury or destructive inflammation of the small vessels, or alveolar wall capillaries (necrotic pulmonary capillaritis).

Hemoptysis

**Diffuse alveolar
infiltrates**



Low hematocrit

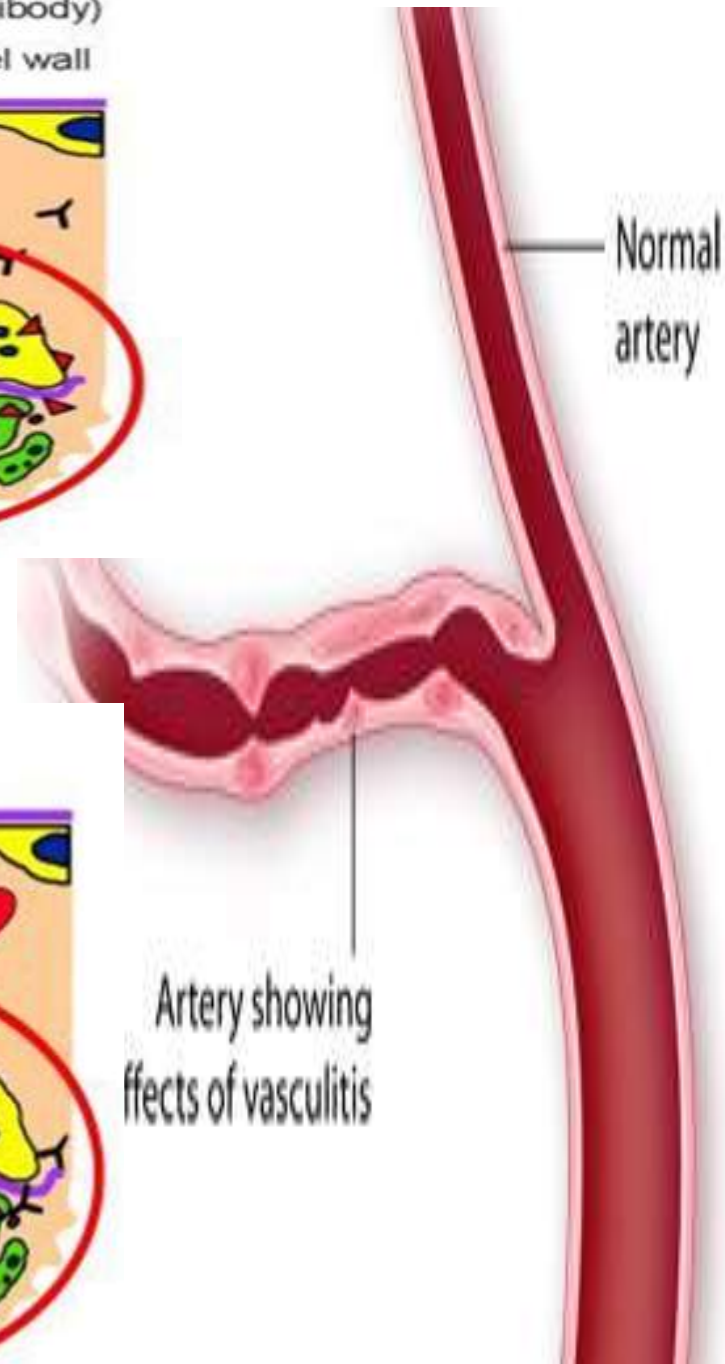
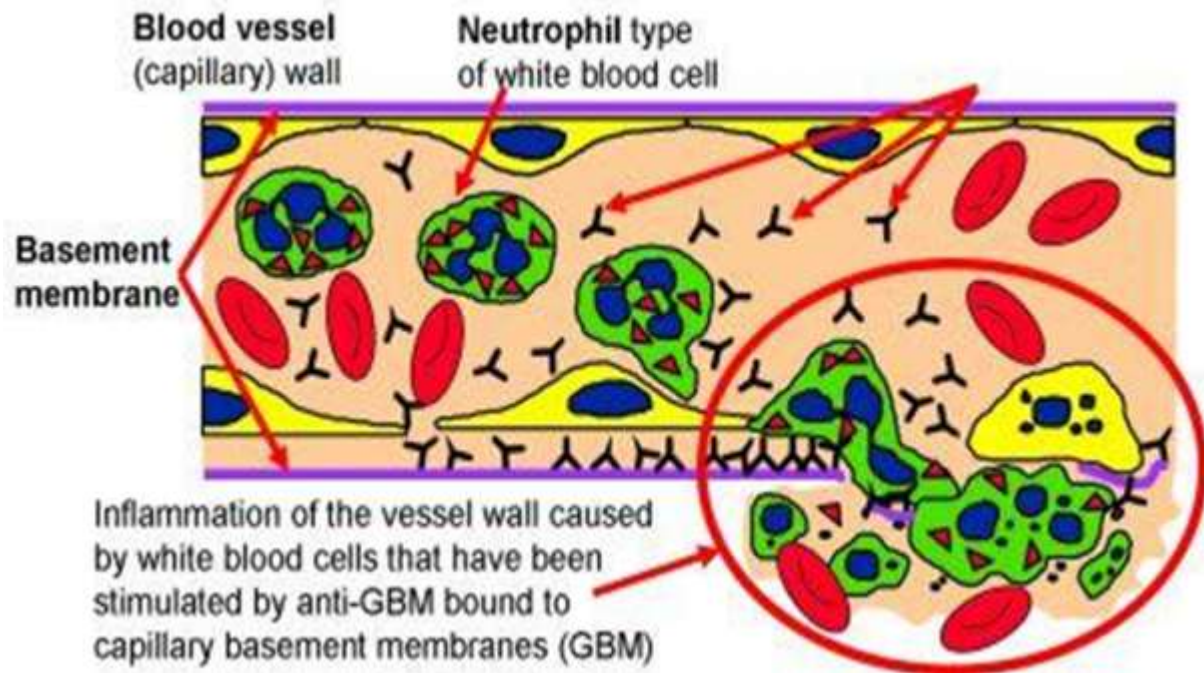
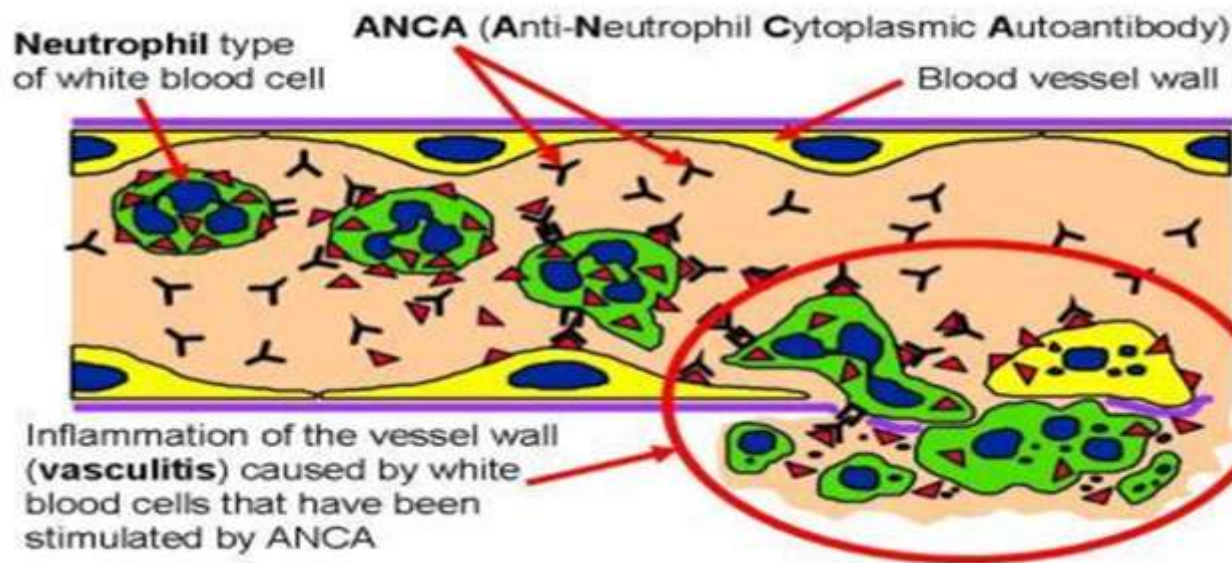
2- RPGM

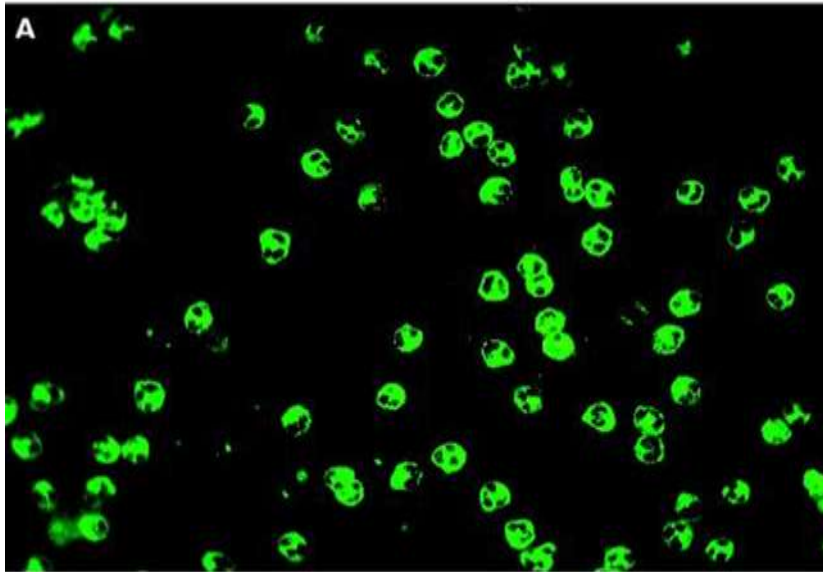
- **The underlying renal pathology in the majority of cases is a form of focal proliferative glomerulonephritis.**

Aetiological / Classification

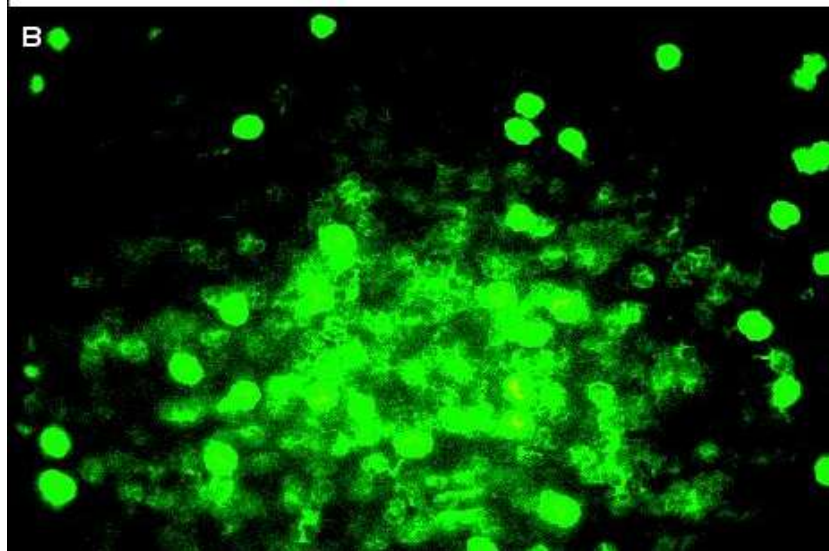


- **ANCA positive vasculitis**
 - Granulomatosis with polyangitis (GPA)
 - Micropolyangitis (MPA)
 - Churg -Strauss syndrome.
- **ANCA negative vasculitis**
 - Behcet,s syndrome
 - IGA nephropathy
 - Henoch schonelin purpra
 - Mixed cryoglobulinemia
- **Antiglomerular basement membrane antibodies**
 - Goodpasture,s syndrome
 - Autoimmune connective tissues diseases
 - SLE & Polymyositis & Scleroderma
- **Drug indused**
 - Hydralazin & Propylthioyoracil & D-Penicellamin
- **Idiopathic PRS.**





C-ANCA



P-ANCA

- **Granulomatosis with polyangitis (GPA)**
- **Micropolyangitis (MPA)**
- **Churg -Strauss syndrome.**
- **Goodpasture,s syndrome**
- **SLE**
- **Behcet,s syndrome**





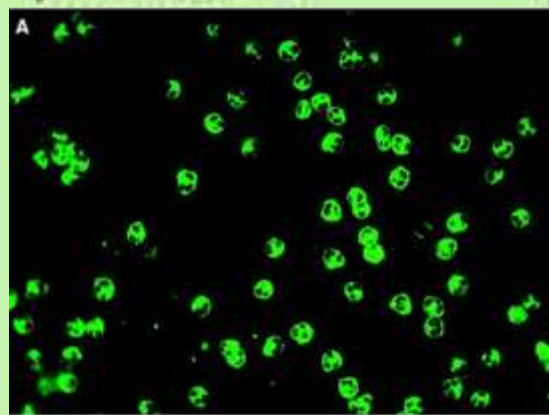
DIAGNOSIS

**Clinical
Radiological
Laboratory
Bronchoscopy
Biopsy**

Wegener's Granulomatosis

Wegener's is infamous for its subtle presentation, and its lethality if it is not correctly diagnosed and treated.

It is caused by autoantibodies against proteinase 3.



Positive c-ANCA
(Anti-neutrophil
cytoplasm Test)



Granulomas &
patchy necrosis
in arteries &
veins

- *Sore Eye
- *Sore Ear
- *Stuffy Nose



*Destruction
of the Face

*Abnormal
Chest Xray

*Lung Cavities
& Bleeding

*Permanent
Kidney Damage
& Failure

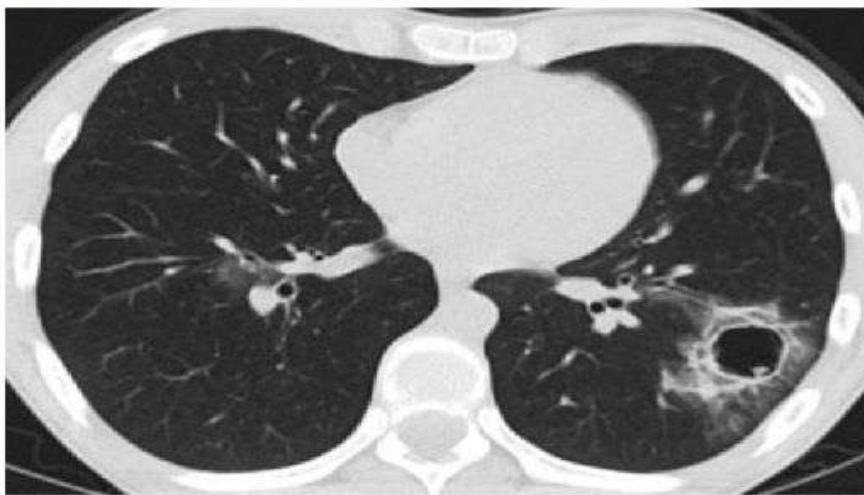
*Sore Joint

*Trace of
blood in
urine

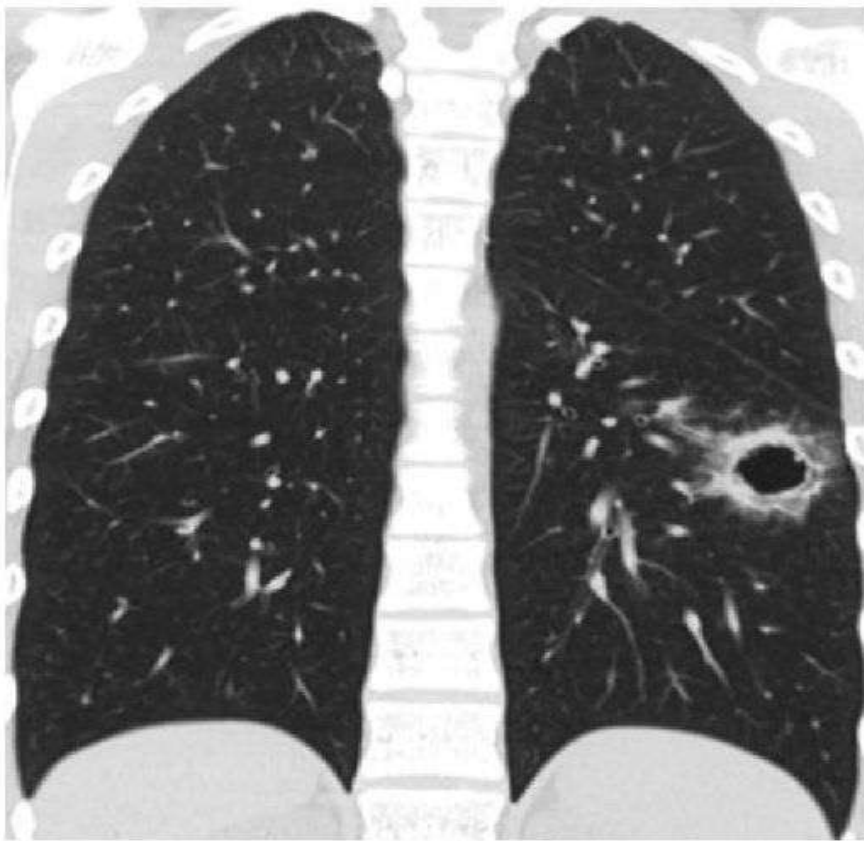
*Gangrene



A



B

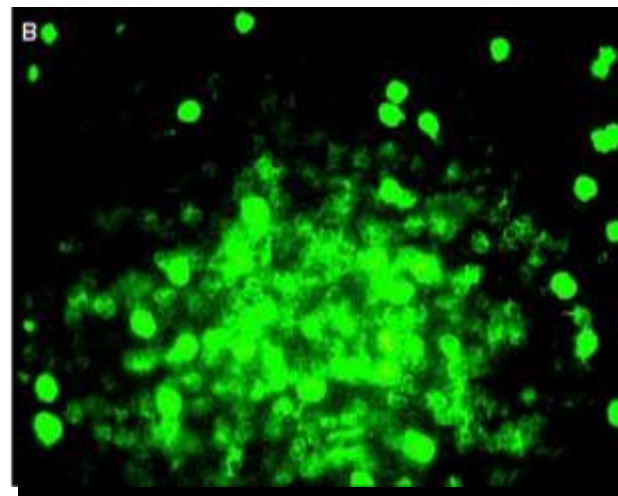


Microscopic Polyangiitis

(Small vessel polyarteritis)

Easy to diagnose and treat -- if you think of it.

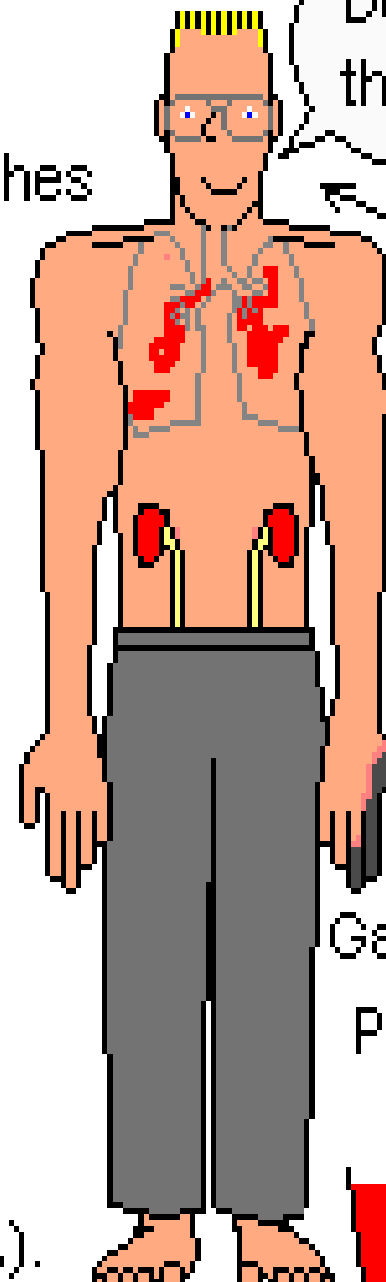
Smaller veins and arteries show patchy 3-layer inflammation



Positive anti-neutrophil cytoplasm test (p-ANCA).

Vague aches and pains

Hemoptysis & infiltrates



Don't miss this either!

Stroke

Heart attack

Bowel infarcts

Nephritis / kidney failure

Gangrene

Peripheral nerve damage

hematuria



Churg-Strauss Syndrome

- Asthma
- Eosinophilia >10%
- Neuropathy
- Pulmonary infiltrates
- Paranasal sinus abnormality
- Extravascular eosinophil infiltration on biopsy

^aThe presence of at least four of the six criteria indicates that Churg–Strauss syndrome is very likely to be the correct diagnosis.



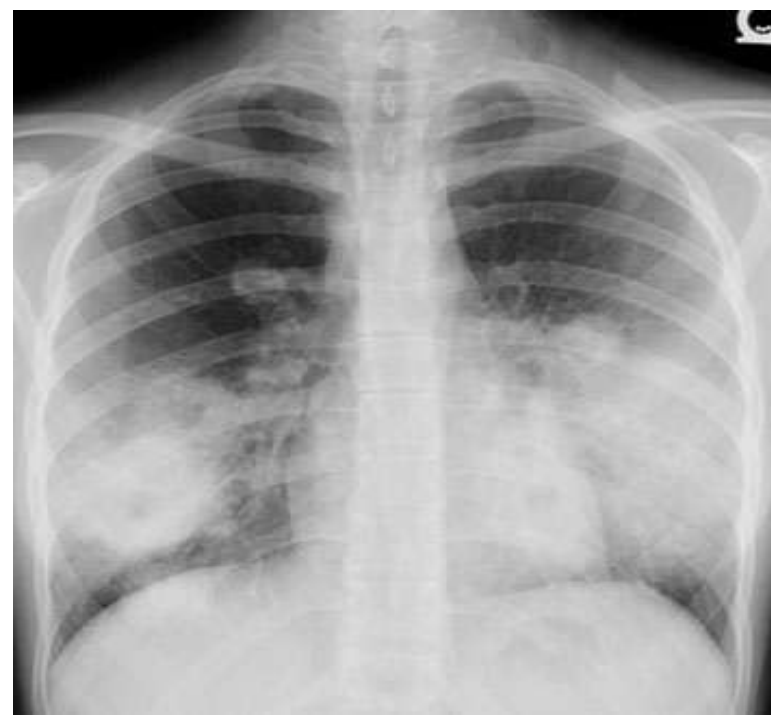
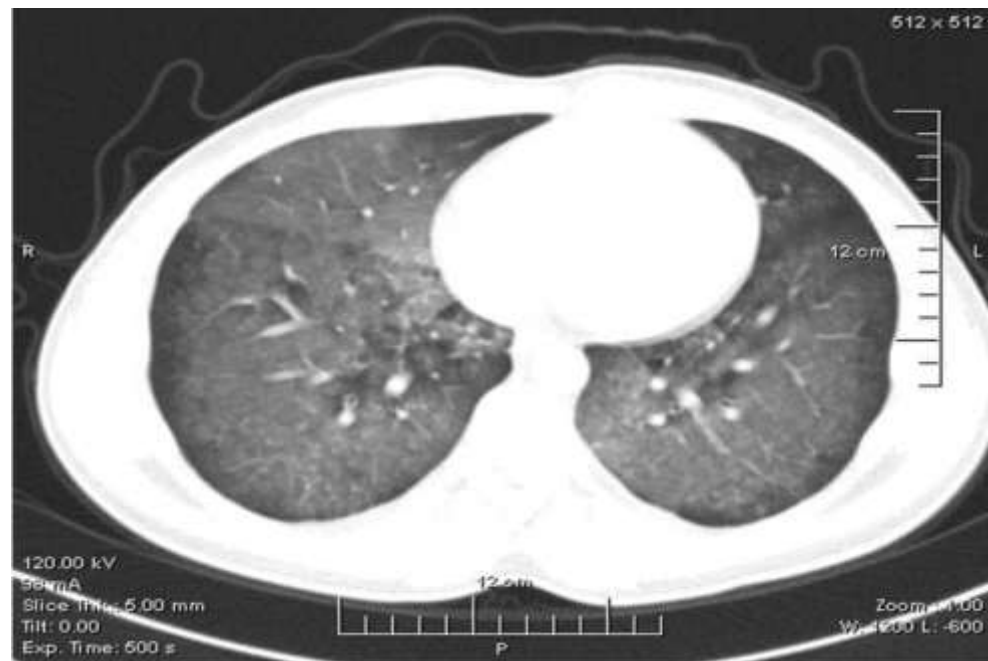
Chest X-ray

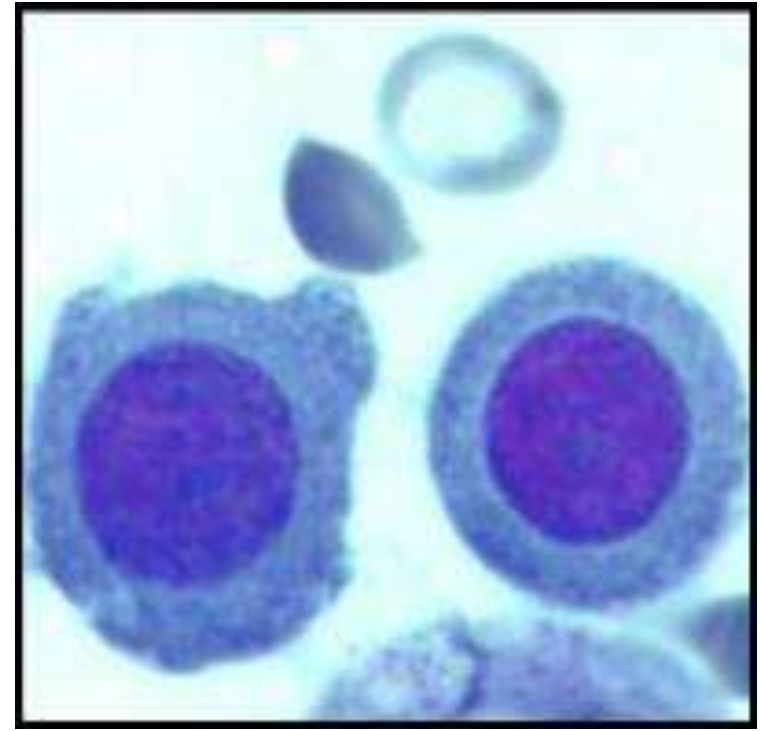
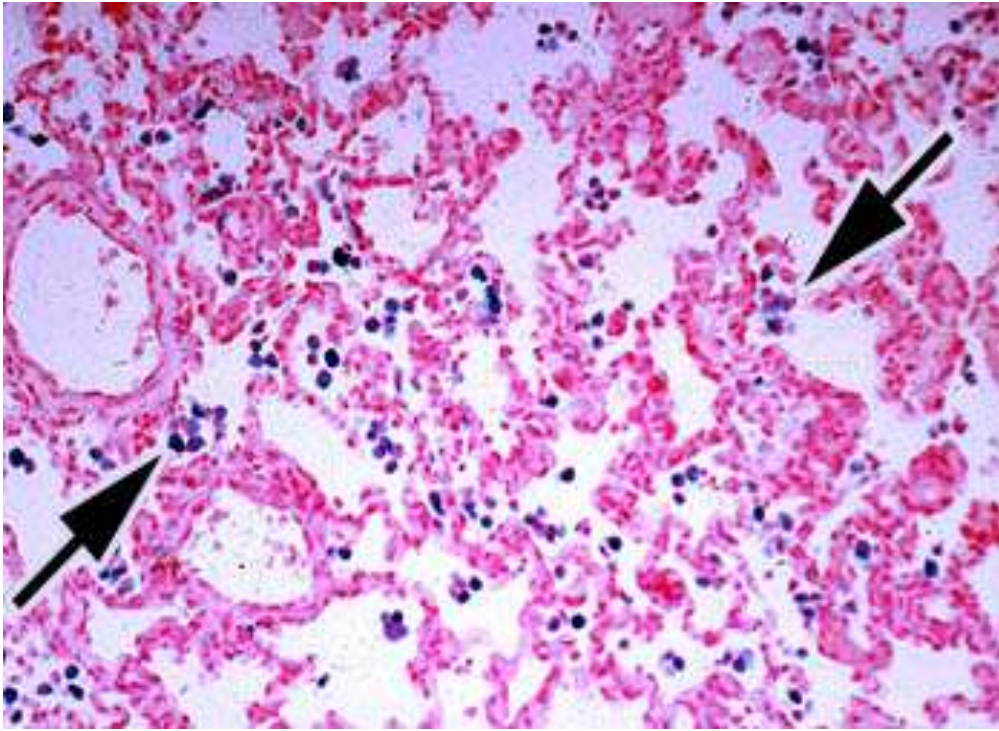


Good pasture,s syndrome

□ Pulmonary manifestations

- The onset of pulmonary hemorrhage may be insidious, with symptoms such as anemia, pallor, weakness, lethargy, dyspnea upon exertion, and, sometimes, dry cough
 - In some cases, onset is acute and includes fever, massive hemoptysis, and acute respiratory failure.
 - In many cases, the symptoms may be present intermittently for weeks to months before the diagnosis is established
 - Only bilateral fine crepitations on local examination
-

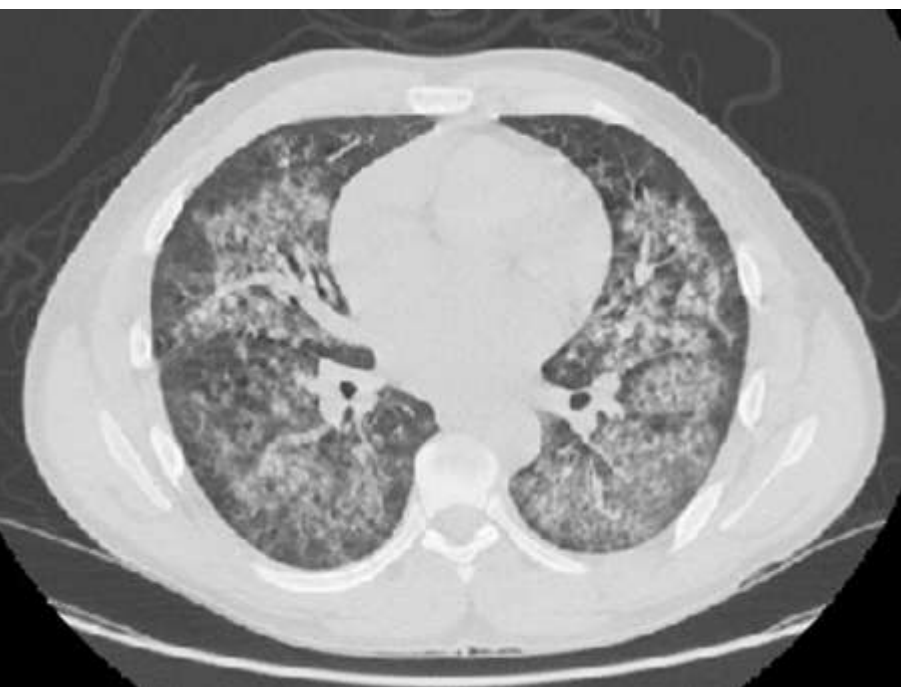
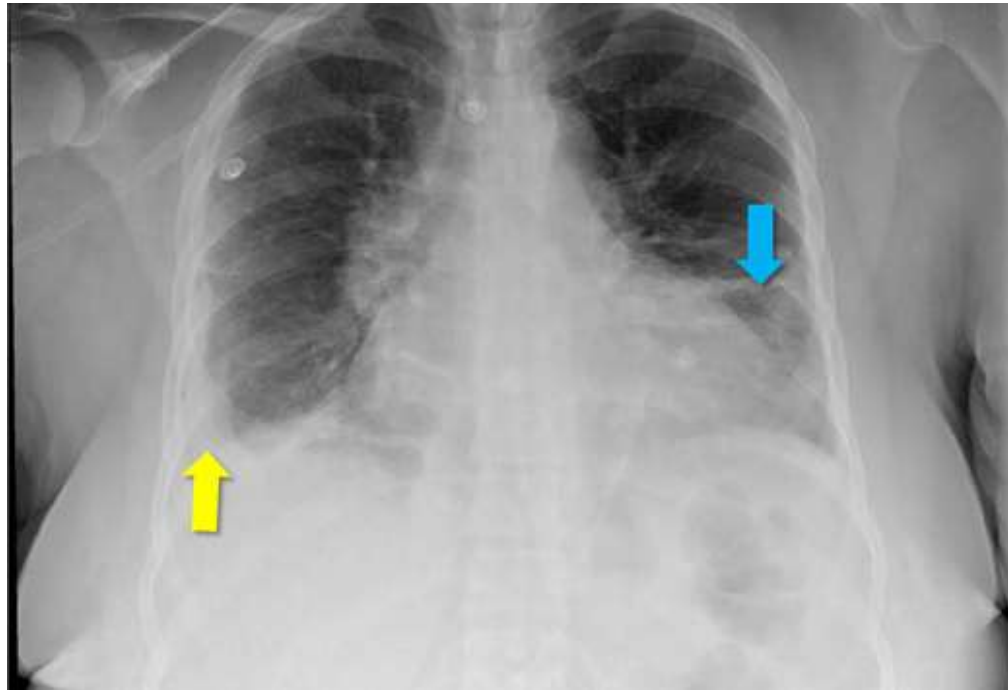




- *Brochoscopy* with BAL and the microscopic detection of **siderophages**.

Lupus lung

- **Upper airway** : VCD & Subglottic stenosis & laryngeal oedema
- **Lower airway** : Small airway disease.
- **Pleura** : Pleurisy & floating effusion
- **Vasculature** : PE & VOD & PHTN
- **Muscles** : Diaphragmatic weakness.
- **Lung parenchyma** :
 - Alveolar hemorrhage
 - IPF
 - Pneumonitis & Atelectasis (shrinkage lung syndrome)
 - Pulmonary oedema



Therapeutic considerations

- The pharmacologic treatment of PRS carry significant risk for drug-associated adverse effects.
- The intensity of the immunosuppressive regimen should be based on disease activity
- Treatment includes two phases
 - Induction phase for achieving remission
 - Maintenance phase for keeping remission
- Treatment monitoring
 - Disease specific and complications
 - Drug specific , side effects and toxicity

.

EUVAS triaging

- (1) limited
- (2) Early, generalized
- (3) Generalized active
- (4) Severe
- (5) Refractory
- (6) Remission

EUVAS Classification	Clinical Features	Five Factor Score	Treatment Options
Limited	Isolated upper airway disease	0	Corticosteroids or methotrexate or azathioprine
Early generalized	End-organ involvement that lacks a clear or immediate threat to organ function. Examples include glomerulonephritis with serum creatinine < 1.4 mg/dl or the presence of minimally symptomatic pulmonary nodules. Constitutional symptoms are common	0–1	Cyclophosphamide + corticosteroids or methotrexate + corticosteroids (for MPA may also consider mycophenolate + corticosteroids)
Generalized active	End-organ involvement with clinically significant impairment of organ function. Examples include glomerulonephritis with serum creatinine > 1.4 mg/dl but < 5.7 mg/dl or pulmonary infiltrates with cough, dyspnea, and impaired exercise tolerance	1–2	Rituximab + corticosteroids or cyclophosphamide + corticosteroids
Severe	Immediate threat of organ failure or death. Examples include severe renal disease with serum creatinine > 5.7 mg/dl, alveolar hemorrhage, and heart failure/cardiomyopathy	≥2	Plasmapheresis + corticosteroids + cyclophosphamide (or rituximab)
Refractory	Disease that has failed to respond to conventional therapy	N/A	Referral to a center of specialized expertise. Consider investigational agents
Remission (maintenance)	No evidence of ongoing vasculitic activity (BVAS = 0)	N/A	If induced with cyclophosphamide then azathioprine ± low-dose oral corticosteroids or methotrexate ± low-dose oral corticosteroids If induced with rituximab no additional maintenance therapy may be required or may use low-dose oral corticosteroids alone

Novel agents for the treatment of pulmonary–renal syndrome [102-113]

Biological agent	Mechanism of action	Indication-study population
Etanercept	TNF α inhibitor	Maintenance therapy in Wegener's granulomatosis
Infliximab	TNF α inhibitor	ANCA-associated vasculitis
Rituximab	Anti-CD20 antibody for B lymphocytes	ANCA-associated vasculitis, refractory to or contraindication to treatment
Mycofenolate mofetil	Suppressor of B lymphocytes and T lymphocytes	ANCA-associated vasculitis, remission maintenance
Leflunomide	Suppressor of T cells	Wegener's granulomatosis, remission maintenance
Antithymocyte globulin	Suppressor of T cells	Severe refractory Wegener's granulomatosis

PRS IN ICU



1. Admitted when there are:

Hemodynamic instability

Severe respiratory distress

Refractory hypoxemia

2- Aiming to :

Minimizing the risk of sepsis

Respiratory and airway management

Cardiovascular and renal support

Minimizing the risk of sepsis

- Patients with PRS frequently die of sepsis (75 %)
- The risk of nosocomial infection in these patients is very high due to immune-suppression.
- Severe infection due to cyclophosphamide occurs in about 10% of cases and has a high mortality.
- Antibiotics covering mainly staph and g -v bacilli
- Antiviral and antifungal should be considered.
- Careful monitoring for bone marrow suppression is indicated.
- **Septtrin prophylaxis** against PCP infection is often used.

Respiratory and airway management

- In **GPA** there may be **subglottic stenosis** which can result in **difficult intubation**. Smaller endotracheal tubes or tracheostomy may be needed.
- In **ARDS** due to diffuse alveolar haemorrhage large tidal volumes or pressure changes may exacerbate damage to pulmonary microvasculature.
- **Lung protective ventilation**, as used in the management of ARDS, with tidal volumes of 6 ml/kg and inspiratory plateau pressures below 30 cmH₂O with permissive hypercapnia may reduce lung injury.

Cardiovascular management

- Patients with pulmonary-renal syndrome may be **hypotensive** because of a combination of
 - Dehydration
 - Haemorrhage
 - Systemic inflammatory response
- Levofed in septic shock keep eye upon CVP.

Renal management

- Many patients develop severe acute renal failure and require **haemodialysis** in ICU.
- Of these the majority will eventually progress to end stage renal failure and require **long term renal replacement therapy**.



- Pulmonary Renal syndromes can be fatal
- In patient with pulmonary infiltrate , look outside cage and ask about his kidney
- In Patient With Renal Disorder Having Dyspnea Don't Ignore His Lung .
- Refractory hypoxemia and or hypotension are an absolute indication for ICU in PRS
- In difficult cases with vague symptoms, sometimes an early renal biopsy can make all the difference

**pulmonary renal syndrome
or
renal pulmonary syndrome**

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WORK TOGETHER





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Have a good time



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